WHAT IS CLAIMED IS:

1. A method of treating a patient with Pompe's disease, comprising: administering to the patient a therapeutically effective amount of human acid alpha glucosidase.

2. The method of claim 1, wherein the patient is administered at least 10 mg/kg body weight per week.

3. The method of claim 1, wherein the patient is administered at least 60 mg/kg body weight per week.

4. The method of claim 1, wherein the patient is administered at least 120 mg/kg body weight per week.

5. The method of any of claims 1-4, wherein the patient is administered a single dosage of alpha-glucosidase per week.

6. The method of any of claim 1-4, wherein the patient is administered three dosages of alpha-glucosidase per week.

7. The method of any of claims 1-4, wherein the amount is administered per week for a period of at least 24 weeks.

8. The method of claim 1, wherein the alpha-glucosidase is administered intravenously.

9. The method of claim 1, wherein the alpha-glucosidase was produced in milk of a transgenic mammal.

10. The method of claim 1, wherein the patient has infantile Pompe's disease.

11. The method of claim 10, wherein the patient survives to be at least one year old.

12. The method of claim 1, wherein the patient has juvenile Pompe's disease.

~~13. The method of claim 1, wherein the patient has adult Pompe's disease.~~

14. The method of claim 1, wherein the alpha-glucosidase is predominantly in a 110 kD form.

15. The method of claim 1, further comprising monitoring a level of human acid alpha glucosidase in the patient.

16. The method of claim 15, further comprising administering a second dosage of human acid alpha glucosidase if the level of alpha-glucosidase falls below a threshold value in the patient.

17. The method of claim 1, wherein the human alpha glucosidase is administered intravenously and the rate of administration increases during the period of administration.

18. The method of claim 17, wherein the rate of administration increases by at least a factor of ten during the period of administration.

19. The method of claim 17, wherein the rate of administration increases by at least a factor of ten within a period of five hours.

20. The method of claim 17, wherein the patient is administered a series of at least four dosages, each dosage at a higher strength than the previous dosage.

21. The method of claim 20, wherein the dosages are a first dosage of 0.03-3 mg/kg/hr, a second dosage of 0.3-12 mg/kg/hr, a third dosage of 1-30 mg/kg/hr and a fourth dosage of 2-60 mg/kg/hr.

22. The method of claim 21, wherein the dosages are a first dosage of 0.1-1 mg/kg/hr, a second dosage of 1-4 mg/kg/hr, a third dosage of 3-10 mg/kg/hr and a fourth dosage of 6-20 mg/kg/hr.

23. The method of claim 22, wherein the dosages are a first dosage of 0.25-4 mg/kg/hr, a second dosage of 0.9-1.4 mg/kg/hr, a third dosage of 3.6-5.7 mg/kg/hr and a fourth dosage of 7.2-11.3 mg/kg/hr.

24. The method of claim 23, wherein the dosages are a first dosage of 0.3 mg/kg/hr, a second dosage of 1 mg/kg/hr, a third dosage of 4 mg/kg/hr and a fourth dosage of 12 mg/kg/hr.

25. The method of any of claims 20-24, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.

26. The method of any of claims 20-24, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, 1hr, 0.5 hr and 3 hr respectively.

27. A pharmaceutical composition comprising human acid alpha .glucosidase, human serum albumin, and a sugar in a physiologically acceptable buffer in sterile form.

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28. The pharmaceutical composition of claim 17 comprising human acid alpha glucosidase, human serum albumin, and glucose in sodium phosphate buffer.

29. A pharmaceutical composition comprising alpha glucosidase, mannitol and sucrose in an aqueous solution.

30. The pharmaceutical composition of claim 27, wherein the sugar comprises mannitol and sucrose and the concentration of mannitol is 1-3% w/w of the aqueous solution and the concentration of sucrose is 0.1 to 1% w/w of the aqueous solution.

31. The pharmaceutical composition of claim 27, wherein the concentration of mannitol is 2% w/w and the concentration of sucrose is 0.5% w/w.

32. A lyophilized composition produced by lyophilizing a pharmaceutical composition comprising human acid glucosidase, mannitol and sucrose in aqueous solution.

33. A pharmaceutical composition prepared by lyophilizing a first composition comprising human acid alpha-glucosidase, mannitol, sucrose and an aqueous solution to produce a second composition; and reconstituting the lyophilized composition in saline to produce a third composition.

34. The pharmaceutical composition of claim 33, wherein the human acid alpha-glucosidase is at 5 mg/ml in both the first and third composition, the mannitol is at 2 mg/ml in the first composition, the sucrose is at 0.5 mg/ml in the first composition, and the saline used in the reconstituting step is 0.9% w/w.

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